Rare Case of Renal Tumour Presenting as Haemoperitoneum- A Case Report

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INTRODUCTION

Classic renal angiomyolipoma (AML) is a benign mesenchymal tumour containing fat, smooth muscle cells and thick-walled blood vessels.[1] However, renal epithelioid angiomyolipoma is considered as a potentially malignant neoplasm. It is a rare tumour constituting 4.6-4.8% of all resected AML. It is characterized by proliferation of predominantly epithelioid cells with approximately one third experiencing metastases.[2] Epithelioid angiomyolipoma (EAML) is included in the family of perivascular epitheloid cell tumour (PECT)which is associated genetically with tuberous sclerosis complex.[3]

PRESENTATION OF CASE

Our patient is an 81-year-old lady who presented with right sided abdominal pain. Ultrasound showed a ruptured renal mass. CECT Abdomen with Urographic Sequence showed a heterogenous enhancing area in the interpolar region. Right Radical Nephrectomy was done immediately and specimen was sent to us for histopathological examination. Intraoperatively a ruptured right renal mass with retroperitoneal hematoma and hemoperitoneum was seen.

Gross and Microscopic Appearance

Kidney had a dark brown haemorrhagic appearance with adherent capsule with areas showing rupture. Cut section showed a haemorrhagic lesion measuring 10 x 9 x 5 cms arising from renal cortex. Microscopy showed a neoplasm with extensive areas of haemorrhage showed cellular areas composed of epithelioid cells showing mild to moderate atypia, intranuclear inclusions with abundant granular eosinophilic cytoplasm. Occasional mitotic figures <2/50 HPF noted. The cellular areas were admixed with adipose tissue, smooth muscle tissue and numerous thick-walled blood vessels. No infiltration, necrosis or renal vessel invasion was seen.

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Figure A. Cut Section of Kidney Showing a Haemorrhagic Lesion Measuring 10 x 9 x 5 cms Arising from Renal Cortex

Figure B. Neoplasm Showing Extensive Areas of Haemorrhage

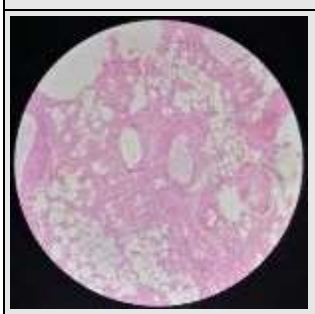

Figure C. Cellular Areas Admixed with Adipose Tissue, Smooth Muscle Tissue and Numerous Thick Walled Blood Vessels

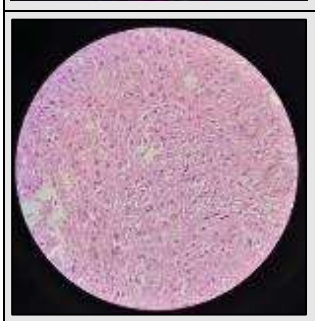

Figure D. Neoplasms Composed of Epithelioid Cells Showing Mild to Moderate Atypia, Intranuclear Inclusions with Abundant Intranuclear Inclusions

DIFFERENTIAL DIAGNOSIS

Oncocytic and clear cell variants of EAMLs can be easily misinterpreted as RCC, renal oncocytoma, or adrenal cortical neoplasm in the renal and adrenal region. Other tumors that enter in the differential diagnosis of EAML are primary or metastatic malignant melanoma, epithelioid smooth muscle tumor, epithelioid peripheral nerve sheath tumor, epithelioid gastrointestinal stromal tumor (GIST), hepatoblastoma, and HCC.

PATHOLOGICAL DISCUSSION

Perivascular epithelioid cell tumour (PECT) represents a family of tumours including classic AML and clear epithelioid cell tumours reported under a variety of names such as EAML and clear cell sugar cell tumour.[4] It belongs to microphthalmia associated transcription factor (MiTF) family of tumours. Mean age of presentation is 50 years. Other sites of involvement are liver, adrenal gland, retroperitoneal soft tissue, urinary bladder, nasal cavity, skin etc., In 2004, WHO considered AML as epithelioid when epithelial morphology is predominant (>80%).[2] Morphologic and genetic studies have shown that renal AML is closely related to the PEComa tumour family, as well as with the tuberous sclerosis complex (TSC) by demonstrating loss of heterozygosity of the TSC2 locus on chromosome 16p. Renal AMLs can occur sporadically or in association with tuberous sclerosis complex. 20% of AMLs are associated with tuberous sclerosis complex, whereas 50% of patients with tuberous sclerosis complex have AMLs, which tend to be multiple and bilateral. The PEComa family includes renal and hepatic AMLs, lymphangiomyomatosis, clear cell 'sugar' tumour of the lung, and a group of similar lesions seen at other sites. Cysts and multiple AMLs are the most common renal manifestation in TSC.[5,6]

In our case, the epithelioid component represents >80% of the tumour size. It was not associated with any syndromes. Literature shows recurrence in about 17% of patients and metastases in about 30% of patients.[7] Our patient has not come with recurrence or metastases. Most common metastatic sites are lymph nodes, liver, lung and peritoneum. TFE3 gene rearrangement is uncommon. Epithelioid angiomyolipoma represents a distinct diagnostic entity and may occur at diverse visceral and somatic soft tissue sites. It mimics morphologically a variety of neoplasms such as RCC, renal oncocytoma, adrenal cortical neoplasm, epithelioid smooth muscle tumour, epithelioid peripheral nerve sheath tumour, epithelioid GIST, epithelioid melanoma, hepatoblastoma, and HCC. This differential diagnosis can be particularly problematic in small biopsies and sometimes even in surgical specimens. Morphologic clues to diagnosis such as islands of mature fat and abnormal vessels should be diligently looked for in surgical specimens. A correct diagnosis of EAML may require prudent use of immunohistochemistry. Due to the variation in immunohistochemical phenotype, ultrastructural analysis may occasionally be of help to establish the diagnosis. The recognition of EAML is important because it is also considered a potentially malignant neoplasm that harbours the potential for metastatic behaviour, particularly in those cases showing infiltrative growth pattern, marked nuclear atypia, necrosis, mitotic activity greater than 1 per 50 high-power fields, and larger (>5 cm) tumour size, but the absence of these features does not always exclude malignant behaviour.[8,9]

Immunohistochemistry includes HMB 45, Melan 1, Microphthalmia Transcription Factor and Cathepsin K.

FINAL DIAGNOSIS

EAMLs have a significant overlap in morphology with more commonly occurring renal neoplasms. EAML is a tumour with malignant potential and can behave aggressively and even metastasize. Here we report a case of a renal tumour

presenting as haemoperitoneum. Once diagnosed, active treatment including radical nephrectomy, chemotherapy and molecular-targeted drugs should be considered and patients should be closely followed.

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